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Case Report

Efficacy of manual lymph drainage in systemic sclerosis: A case report

Manual lymph drainage in individuals with systemic sclerosis

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Abstract
The aim of this case report was to evaluate the efficacy of manual lymph drainage (MLD) in a patient with Systemic Sclerosis in relation to pain, dyspnea, insomnia, edema, skin involvement, hand functions and quality of life . A 57-year-old patient with SSc who applied manual lymph drainage is presented in this case report. A Visual Analog Scale (VAS) was used to assess pain, dyspnea, edema and insomnia, volumetric measurements were taken of hand volume, the Hand Mobility in Scleroderma (HAMIS) test was applied for hand function, the Modified Rodnan Score (MRS) for skin involvement and the Short Form-36 (SF36) for Quality of Life. It has been observed that the application of MLD to systemic sclerosis patient has positive effects on skin involvement, edema, pain, dyspnea, insomnia, and quality of life. It is thought that it will be beneficial to add MLD application to the physiotherapy treatment program for patients with systemic sclerosis.

Keywords
Systemic sclerosis; Manual lymph drainage; Edema

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Introduction

Systemic sclerosis (SSc) leads to restrictions because of the hardening of the skin, joint and muscle involvement. Problems on the hands show great variability depending on the stage of the disease. In the early stage, tissue edema is seen, especially on the fingers. Edema affects the face, extremities and trunk. Over time, fibrous tissue develops on the site of edema in the hands and fingers, and this reduces movement and functionality. This then restricts daily living activities and diminishes the quality of life [1,2].

Manual lymph drainage (MLD) is the application of light massage to the skin that increases smooth muscle contraction around the lymphatic vessels and thereby increases lymphatic flow and eliminates edema and excess amounts of interstitial fluid [3,4]. There is also an effect on the autonomic nervous system. Sympathetic responses are reduced and parasympathetic responses are increased. Decreased sympathetic system responses reduce the chronic inflammatory process [4].

MLD is used in patients with upper extremity edema, which has formed following breast cancer treatment. It is also used in the treatment of edema, which has formed for different reasons, and in a very broad spectrum of diseases and symptoms, such as venous and inflammatory edema (after trauma, infection, or surgery), Sudeck disease, scar treatment, rheumatoid diseases, headache and migraine [3].

The complex pathogenesis of edema in SSc includes microvascular changes, increasing sympathetic activity and inflammatory changes. In addition to the known microvascular changes in SSc, impaired lymphatic circulation in the upper extremities has been reported [5]. The presence of finger and hand edema in SSc can be considered to support the therapeutic approach of MLD. But there are not many studies on this subject in literature.

There are few studies in literature related to the effect of MLD on pain. It has been reported that pain is caused by the pressure of edema [2,3,6]. Therefore, knowledge of the changes in the patient's perception of pain during and after the application of MLD in clinical applications would be of benefit in applying these practices with more awareness.

The Health Assessment Questionnaire (HAQ) is correlated with skin involvement, hand deformities, muscle strength and renal or cardiac impairment in SSc. In addition, respiratory problems are seen in SSc [2]. There are not many studies that examined the efficacy of MLD on respiratory problems [7]. The autonomic nervous system maintains homeostasis by the coordinated work of the sympathetic and parasympathetic systems. Parasympathetic system regulates the respiratory function, by providing a smooth muscle bronchoconstrictor effect. According to Vodder methods, MLD techniques can have effects on the autonomic nervous system. It can reduce sympathetic activity and indirectly increase the parasympathetic system. However, respiratory techniques are also used in MLD, Diaphragmatic breathing increases lymph flow and provides fluid flow in the body and repels edema [3,4,7].

Sleep quality decreases in patients with systemic sclerosis due to edema and pain. As a result, the quality of life will be affected in patients with systemic sclerosis due to pain, respiratory problems, edema, skin involvement and decreased hand functions [8]. This case report was planned to evaluate the efficacy of manual lymph drainage (MLD) applied to a patient with Systemic Sclerosis in relation to pain, dyspnea, insomnia, edema, skin involvement, hand functions and quality of life .

Case Report

The patient was 158 cm in height, weighed 82 kg, was a housewife and had no disease other than SSc. A 57-year old female with an 8-year history of SSC was followed up with a medical treatment in the Rheumatology Department of Pamukkale University Medical Faculty. Respiratory rate was 14/min. The facial skin was taut, with limited lip movement and the facial features were unclear. Hands and fingers appeared to be swollen. The patient was in the edematous phase. There were no ulcers on the hands (Figure 1). The patient had no history of surgery and the family history revealed heart disease in her mother. The patient reported that she did not smoke or drink, and had recently experienced sleep problems and shortness of breath when walking and particularly when ascending stairs.

She stated that the swelling and pain have increased, especially in the last month. The patient was admitted to the physiotherapy program with complaints of severe pain in the hands and feet that were restricting daily living activities. No change was made to the medical treatment while the physiotherapy program was applied. Informed consent for publication of the case was obtained from the patient. Evaluations and treatment of the patient were made by different physiotherapists. Before and after treatment, the patient was evaluated in respect of pain, dyspnoea, insomnia, and edema in the hand using a Visual Analog Scale (VAS) on a scale of 0-10 where 0 = the best status and 10 = the worst status [2].

Volumetric measurement to evaluate hand volume: the patient was instructed to place each hand separately into the water as far as the wrist, and the volume of the water displaced was calculated separately for each hand. The measurements were taken twice and the average was calculated for analysis. To prevent diurnal changes in edema, all the volumetric measurements were taken at the same time of day [1].

The Hand Mobility in Scleroderma (HAMIS) test is a valid and reliable tool for the evaluation of hand function in SSc and was applied to both hands for hand functional evaluation. The 9-item performance areas of the HAMIS test include holding and different movements in different dimensions, with tools and movements that are all related to a part of daily living activities. Each exercise is graded on a scale of 0-3, where 0 = normal function and 3= insufficient function. A maximum score of 27 can be obtained for each hand [2].

Quality of life was evaluated with the Short Form -36 (SF36). This is a 36- item scale, under separate subheadings, which is used to measure the functional status of an individual. The 8 sections of the scale are scored from 0 to 100. The subparameters evaluated in the scale are social competence, cognitive health status, physical capability, pain, emotional strength, energy/vitality, general health perception, and physical strength. Each subparameter is scored differently, with higher scores indicating an increased rate of development in the health status [7].

The Modified Rodnan Score (MRS) was used to evaluate skin

Table 1. Pre-treatment and post-treatment evaluation of patient's results

Variables	Pre-Treatment		Post-Treatment		Comparison Pre- and Post-Treatment	
Pain (VAS, cm)	7		4		3	
Dyspnea (VAS, cm)	10		8		2	
Insomnia (VAS, cm)	10		5		5	
Edema (VAS, cm)	9		5		4	
MRS (0-51)	22		14		8	
Variables	**Right**	**Left**	**Right**	**Left**	**Right**	**Left**
Volumetric measurement (cm^3)	640.64	677.60	566.72	542.08	73.92	135.52
Hamis (0-27)	8	5	8	5	0	0
Hamis (0-27)	7.5		23.75		16.25	
SF-36 Physical Function (0-100)	24.875		26.825		1.050	
SF-36 Mental Health (0-100)	0		25		25	
SF-36 Pain (0-100)	0		33.3		33.3	
SF-36 Emotional role (0-100)	12		24		12	
SF-36 Mental well-being (0-100)	0		25		25	
SF-36 Energy Exhaustion (0-100)	20		35		15	
SF-36 General health (0-100)	25		87,5		62.5	
SF-36 Social function (0-100)	22		14		8	

Figure 1. Our patient's hands pre-treatment

Figure 2. Our patient's hands post-treatment

involvement. The MRS is used in the follow-up of the course of the disease, with measurement of the skin by palpation. Scoring is applied as 0: normal skin, 1: mildly thickened skin, 2: moderately thickened skin (cannot be held), and 3: severely thickened skin (cannot be moved). The maximum score is 51, with higher scores indicating higher skin involvement. The first symptoms of skin changes in the early stage of the disease may not be evident, and in the late stage, the skin may be observed with an atrophic and hardened appearance [1].

The therapeutic exercises in the treatment program were applied with MLD using the Vodder technique. The program was applied as 15 sessions, 5 days a week for 3 weeks. The MLD technique was applied for 45 mins in each session. In all the sessions, the applications were started with the deep abdominal technique. First, MLD was applied to the neck region, after starting with effleurage, the swimming in the terminus technique was used. The lymph nodes in the neck were stimulated. Then MLD was applied to the trunk, which was drained in the first five days. In the following days, the axillary and inguinal lymph nodes were stimulated, then MLD was applied to the extremities [3-5]. In the working phase of MLD, a certain pressure is given, then it is moved on to the relaxation phase. These changes in pressure create a pump effect and lymph flow increases. The drainage is performed repeatedly and rhythmically [3,4].

Pre-treatment, the VAS scores of the patient were 7 for pain,10 for respiratory difficulty, 10 for insomnia, and 9 for edema felt in the hand during daily living activities, the MRS score was 22, volumetric measurements were 640.64 cm3 for the right hand and 677.6 cm3 for the left hand (Figure 1), the HAMIS score was 8 for the right hand and 5 for the left hand, and the SF36 subparameter scores were physical function 7.5, mental health 24.875, pain 0, emotional role 0, mental wellbeing 12, energy consumption 0, general health 20, and social function 25 points. Following treatment the evaluation results were VAS scores of 4 for pain, 8 for respiratory difficulty, 5 for insomnia, and 5 for edema felt in the hand during daily living activities, the MRS score was 14, volumetric measurements were 566.75 cm3 for the right hand and 542.08 cm3 for the left hand (Figure 2), the HAMIS score was 8 for the right hand and 5 for the left hand, and the SF36 subparameter scores were physical function 23.75, mental health 26.825, pain 25, emotional role 33.3, mental wellbeing 24, energy consumption 25, general health 35, and social function 87.5 points.

When the post-treatment results were compared with the pre-treatment values, there was an improvements of 3 for pain, 2 for respiratory difficulty, 5 for insomnia, and 4 for edema felt during daily living activities, according to the VAS points, an improvement of 8 points in MRS, volumetric measurements decreased by 73.92 cm3 in the right hand and by 135.52 cm3 in the left hand, and the HAMIS score did not change. An increase was observed in the SF36 subparameters.

Discussion

This case report was planned to evaluate the efficacy of manual lymph drainage applied to the scleroderma patient in the edematous phase on pain, dyspnea, insomnia, edema, hand functions and quality of life. As a result of this study demonstrated that MLD treatment applied to this SSC patient,

positive improvements were obtained in skin involvement and edema in the hands, and this was seen to be reflected in the quality of life.

Difficulties in daily living activities occur because of conditions such as hardness in the hand, the presence of edema pain, reduced grip strength and slower hand movements that develop in connection with systemic sclerosis [1]. In patients with systemic sclerosis, quality of life is impaired due to pain, respiratory problems, edema and hand functions [8]. It was reported that MLD reduced edema in the upper extremities and increased quality of life in a patient with edema due to breast cancer [3]. Similar to the literature, there was a decrease in the edema volume in both hands. MLD stimulates the lymphatic vessels, and contributes to the movement of fluid in the lymphatic vessels and excretion of excess interstitial fluid. It also reduces sympathetic system activity and increases parasympathetic system activity, thus providing vasodilatation of the blood vessels and an improvement in circulation [4]. As a result, the edema is removed from the body through increased circulation.

Previous studies have shown that manual lymph drainage improves quality of life by reducing pain and edema in reflex sympathetic dystrophy, fibromyalgia and psoriatic arthritis. [3,6]. Self-perceived pain in the current patient was reduced following MLD, which was attributed to a decrease in the pressure created by edema, as the skin and subcutaneous nerve fibres are affected by the pressure of edema [4]. In the current patient, self-perceived sleep complaint in VAS was reduced. We think that the reduction in pain and edema also reduce sleep problems.

In addition, as a result of the treatment, the current patient reported a decrease in respiratory problems. Parasympathetic system regulates the respiratory function by providing a smooth muscle bronchoconstrictor effect. By increasing parasympathetic activity, the use of MLD has a positive effect on respiration [7].

It is important to use methods to reduce hand impairment and disability from the early stage of the disease. Because in SSc patients, the hand involvement leads to notable difficulties in daily living activities. It was also found that the quality of life was worsens to a greater extent in patients with skin involvement [8]. In the current patient, a decrease was determined in the MRS, and it is thought that the decreased area of skin involvement had a positive effect on the quality of life.

It was reported that MLD, added to the rehabilitation program, improved hand function and quality of life in SSc patients [2]. A decrease was observed in edema, skin involvement, pain and respiratory and sleep problems in the current patient. An increase was obtained in all the SF36 total point and subparameters. In patients with edema and lymphedema, the application of MLD has been shown to provide a reduction in extremity volume and pain and an increase in quality of life [3]. MLD improves quality of life for a variety of reasons. The pressure on the tissues is reduced and the lymphatic transport capacity is increased, which reduces the volume of the affected body part. In the same way that the reduced volume can reduce discomfort (pain, tension, weight), the functionality of the affected body area is increased. In addition to the effect on lymphatic vessels, blood flow in superficial arteries and collecting vessels increases and wound healing is accelerated. Moreover, MLD should be combined with exercise and skin care, which have a positive effect on the quality of life [1-4]. Quality of life is thought to have been positively affected because of the reduction in edema volume, skin involvement, pain, and respiratory and sleep problems.

They reported that the reduction was obtained in the HAMIS test in SSc patients. MLD with other treatments was applied in this study [2]. But in current patient, no change was determined in the HAMIS test. Combining manual lymph drainage with other treatments may increase its effectiveness. However, we think that our study is related to its short duration. Greater improvements could be obtained in the long term.

Previous studies on rehabilitation in SSc have included small patient samples and sometimes no control group. Although rehabilitation has been shown to be considerably effective in treatment, there is no evidence as yet that it can actually halt the disease [1,2]. High-quality randomized controlled studies with a longer follow-up are needed to prove the efficacy of MLD in SSc. However, large rehabilitation RCTs are difficult to conduct due to the SSc patients lack of information about the use and the efficacy of MLD and rehabilitation, the scarcity of physiotherapists specialized in MLD in evaluating and treating rheumatic patients, the difficulty of patients in accessing rehabilitation centers, and as patients are not in the edematous phase.

The effect of rehabilitation diminishes over time after treatment ends. Therefore, it should be emphasized that the treatment of SSc patients should be applied regularly and continuously. The limitations inherent in case studies are difficult to generalize as there is a single patient. The lack of objective evaluation of respiration before and after treatment can be considered a limitation of this study. In addition to our study is the short duration of treatment (3 weeks). After the treatment, how long the effect of the treatment continues was not followed. However, the observed result is consistent with the currently available literature.

In conclusion, the application of MLD treatment to a patient with scleroderma had positive effects on skin involvement, edema, pain, respiration, and quality of life. In SSc, MLD, when performed by experienced physiotherapists is safe and does not cause significant adverse effects for SSc patients. It will be beneficial to add MLD application to the physiotherapy treatment program in patients with systemic sclerosis. However, there is a need for further randomized, controlled studies to better reveal the efficacy of MLD in these patients.

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Scientific Responsibility Statement
The authors declare that they are responsible for the article's scientific content including study design, data collection, analysis and interpretation, writing, some of the main line, or all of the preparation and scientific review of the contents and approval of the final version of the article.

Animal and human rights statement
All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. No animal or human studies were carried out by the authors for this

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Conflict of interest

None of the authors received any type of financial support that could be considered potential conflict of interest regarding the manuscript or its submission.

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